Oral Oncology EXTRA (2006) **42**, 82–84

ELSEVIER

ORAL ONCOLOGY EXTRA

http://intl.elsevierhealth.com/journal/ooex

CASE REPORT

Papillary cystadenocarcinoma of the submandibular gland

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Received 7 September 2005; accepted 11 September 2005

KEYWORDS
Papillary cystadenocarcinoma;
Salivary gland neoplasm

Summary Papillary cystadenocarcinoma is an extremely rare malignant neoplasm characterized by cysts and papillary endophytic projections. It was first defined in 1991 by WHO. We presented a case of papillary cystadenocarinoma arising from the submandibular gland in a 78-year-old male patient, and discussed the clinical, histopathological and treatment features of this rare entity in the light of the current literature.

Introduction

Papillary cystadenocarcinoma is an extremely rare malignant neoplasm that was first defined in 1991 by WHO.[4] It has also been called malignant papillary cystadenoma, low-grade papillary adenocarcinoma, or mucus-producing adenopapillary carcinoma.[2] In this article, we report an unusual case of papillary cystadenocarcinoma arising from the submandibular gland.

Case report

A 78-year-old male patient complaining of painless mass in the right upper neck was referred to our department. On examination, a solid 5×3 cm mass was palpated in the right submandibular region. The rest of the ear–nose–throat examination was normal. The patient has had a history of myelofibrosis for 6 months and asthma for many years. Then fine needle aspiration biopsy was performed under ultrasonographic view and revealed 'malignant epithelial cells'. A computed tomography revealed a mass lesion in the right submandibular gland without any involvement of the surrounding tissues and cervical lymph nodes. The patient underwent right total submandibular gland extirpation under general anesthesia. Macroscopically,

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1741-9409/$ - see front matter
doi:10.1016/j.ooe.2005.09.005

anterior half of the gland was replaced by a cystic multilobulated tumor mass and measured $2.5 \times 2.5 \times 2$ cm in size (Fig. 1). Microscopically, it was well separated from the healthy salivary tissue, and predominantly composed of cystic areas. Many of these cystic areas were occupied by papillary endophytic projections (Fig. 2). In some areas, there were round-oval epithelial cell components with hypercromatic nuclei, and mucus secreting cells (Fig. 3). Mitotic figures were rare. No psammoma bodies, vascular or perineural invasion were identified. Histolopathologically, this primary tumor was considered as a low-grade papillary cystadenocarcinoma.

He has been followed up regularly at our department for 14 months, and no regional recurrence or distant metastases were noted.

Figure 1 Macroscopic view of the mass. Note the multiloculated portion of the tumor.

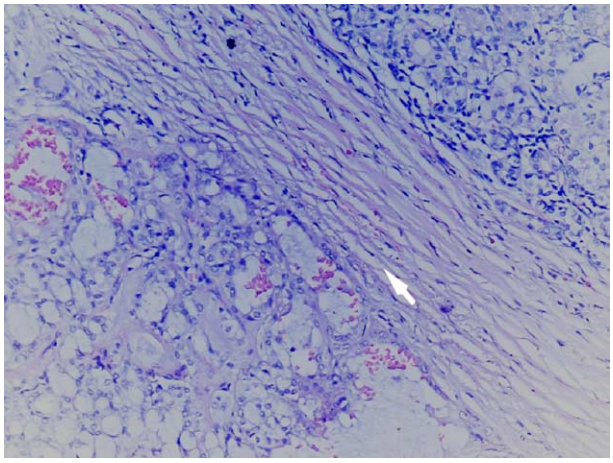

Figure 2 Arrow indicates the cystic portion of the tumor which was filled with papillary projections of the atypical cells (Hematoxylin—Eosin ×10).

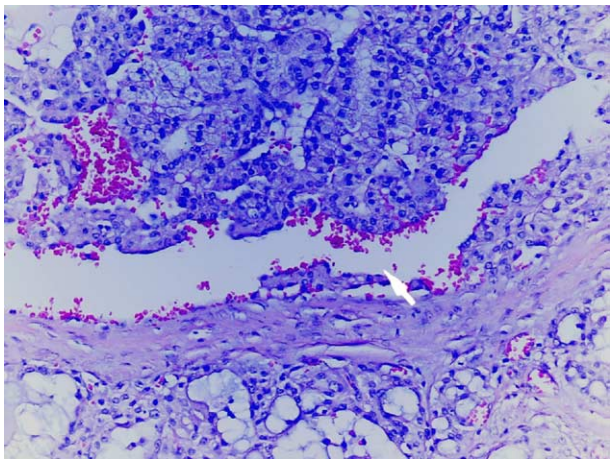

Figure 3 Arrow shows the round-oval epithelial cell components with hypercromatic nuclei, and mucus secreting cells. As noted, mitotic figures are rare (Hematoxylin—Eosin ×20).

Discussion

Papillary cystadenocarcinoma is defined by WHO as a low-grade malignant tumor characterized by cysts and papillary endophytic projections. The epithelium is cuboidal or low columnar, mucus secreting cells can also be present, and malignancy is confirmed by nuclear pleomorphism, mitosis and infiltrative growth pattern.[3] Most of the patients present a slowly growing and asymptomatic mass, as in our case. Despite of its low-grade nature, there have been several reports indicated that papillary cystadenocarcinoma may have a more aggressive behavior, and a higher grade pathological features.[3] This poorly differentiated type of papillary cystadenocarcinoma has a metastatic potential to cervical lymph nodes. However, there are no defined parameters for grading in papillary cystadenocarcinoma due to lack of well-defined case series. In our case, the histopathological features, operation findings, and post-operative follow-up were consisted with the diagnosis of low-grade papillary cystadenocarcinoma.

Immunohistochemical features are still controversial, but many reports indicate that CEA, CA 19-9, and CA125 are positively expressed in papillary cystadenocarcinoma. However, we found positive staining for cytokeratin, smooth muscle antigen, and S 100, but no staining for CEA.

According to Foss et al. series of salivary gland cystadenocarcinomas ($n = 57$), it occurs predominately in parotid gland (71%), next in the minor salivary glands (20%), including two cases in the sublingual glands (3.5%). However, they did not report any case in the submandibular gland.[1] Based

on our English literature search; this is probably the first case in the submandibular gland.

The differential diagnosis of papillary cystadenocarcinoma includes adenocarcinoma, salivary duct carcinoma, polymorphous low-grade adenocarcinoma, and metastatic papillary carcinomas.[2,3,5] The present case had no history, signs and symptoms of a primary adenocarcinoma focus in the body.

In conclusion, we report an extremely rare case of papillary cystadenocarcinoma arising from the submandibular gland. Salivary gland cystadenocarcinomas represent a distinct group of malignancies that have an indolent biologic behavior that its exact pathophysiological mechanism is still unresolved.

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